All manuscripts will be prepared with the International Committee of Medical Journal Editors - Uniform requirements for manuscripts submitted to Biomedical Journals, 1993.

Manuscripts submitted for publication are accepted on the understanding that they are contributed exclusively to *The Central African Journal of Medicine*. A statement to that effect should be included in the letter accompanying the manuscript.

Communications concerning editorial matter, advertising, subscriptions, change of address, etc. Should be addressed to the Administrative Manager, P. O. Box A195 Avondale, Harare, Zimbabwe.

The subscription rate for **surface transmission** including postage for year 2001 is Z$770.00 locally; Africa US$160.00 for individuals and US$215.00 for institutions; and US$210.00 for individuals and US$230.00 for institutions for the rest of the world per annum. The subscription rate for **airmail transmission** for year 2001 in Africa is US$275.00 for individuals US$290.00 for institutions and US$70.00 for postage; and US$300.00 for individuals US$320.00 for institutions and US$70.00 for postage for the rest of the world per annum.

Owned and published by the *Central African Journal of Medicine* in conjuction with the Faculty of Medicine

University of Zimbabwe

STRAND MULTIPRINT (PVT) LTD

Profile of the Nigerian sickle cell anaemia patients above 30 years of age

*A YETUNDE, **C C ANYAEGBU

Abstract

Objective: To examine the haematological profiles of patients with sickle cell anaemia above the age of 30 years.
Design: Prospective cross sectional study.
Setting: Department of Haematology University College Hospital, Ibadan, Nigeria.
Subjects: 98 patients with sickle cell anaemia above the age of 30 years.
Main Outcome Measures: Clinical and haematological profiles of the patients.
Results: There were 75 females and 23 males. The steady state packed cell volume was 21 to 28% with a median of 24% Haemoglobin F (HbF) level in 33 patients was greater than 3% but less than 5%. Thirty six patients had a HbF which was greater than 5% but less than 10%. Twenty three patients had a HbF level which was greater than 10% but less than 15%. Five patients had a HbF level greater than 15% and only one patient had a HbF level of 22%.

The severe complications of sickle cell anaemia were not observed in 87 patients (88.8%), though some age related complications like Grade 4 ischaemic necrosis of the femoral head, chronic inflammatory liver changes and ophthalmic complications were observed. Eighty six patients had never had a blood transfusion, while 76 had never been admitted into hospital. Thirty four patients had a mild to moderate degree of splenomegaly while 64 patients had mild to moderate degree of hepatomegaly. Seven of the patients had chronic ulcers which spanned five to 12 years before the study. Five patients, however, had had cerebrovascular accidents. The cohort of patients with HbF level greater than 10% had not experienced priapism, acute chest syndrome, cerebrovascular accidents or chronic leg ulcer. All the patients have had a form of Western education, have a good knowledge of the disease and possible outcome.
Conclusion: The study strongly suggests that for the survival of the sickle cell anaemia patient, educational background of patients and their parents, awareness of hospital treatment and early presentation in hospital are major contributory factors. Also, adequate medical care, maintenance of disciplined life styles, stable or sedentary occupation and good family support, prompt and effective treatment of complications, all contribute to better health and hence a longer life span.

Cent Afr J Med 2001;47(4):108-11

**Department of Haematology*
***Department of Chemical Pathology*
University College Hospital
Ibadan
Nigeria

Correspondence to:
Dr A Yetunde
Department of Haematology
University College Hospital
Ibadan University
Ibadan
Nigeria

Introduction

Haemoglobin S occurs with greatest prevalence in tropical Africa and clinicians have generally associated sickle cell disease with high morbidity and mortality.[1-3]

The heterozygous frequency in Nigeria is usually about 25% but in some areas it reaches 32.5%.[4] Patients with sickle cell anaemia (SCA) constitute about 2 to 3% of the Nigerian population. While the disease appears benign in some patients, it runs a crippling course in others. There are a few reports of patients surviving till the fourth or fifth decade but this is a very rare occurrence in Nigeria. Thus, patients over the age of 30 years form a very small proportion of the patients. It has been implied that inherited and acquired factors influence the pathogenesis and clinical symptoms of the disease.[1] Hence, this results either in death in the early years in some patients or cases discovered late in life as a result of chance survey.[1] With improvement in the living standard and increasing availability of health care, it has been observed that more patients with sickle cell anaemia in Nigeria survive into adolescence and maturity, and they are able to reproduce and some are in gainful employment.

Patients with SCA vary in the haematological and clinical features of their disease.[5] The observed variability of the clinical manifestations amongst SCA patients is thought to be genetically determined hence the genetic regulation of the rate of progression of sickle cell induced vasculopathy and the clinical expression of the disease while most clinical complications are thought to be age specific.[6-8]

Considerable knowledge has accumulated in the understanding of the physiology and pathogenesis of SCA more than in any other inherited disease. Extracellular factors that influence the disease include environmental factors, low oxygen tension, cardiac or respiratory disorders temperature and acidosis, while intra-cellular factors are genetically determined.[7] The failure of the spleen to filter and effectively destroy polysaccharide encapsulated bacteria accounts for the high septicaemic death rate among SCA patients.

It is now clear that recent advances in treatment like the use of Hydroxyurea, and exchange transfusion regimes have enhanced the quality of life and some of these patients now live longer.

Owing to the dearth of information regarding SCA in those aged 30 or above, patients aged 30 years or more were studied and the observations recorded with regard to clinical and laboratory findings are presented.

Materials and Methods

Patients above 30 years of age diagnosed as having SCA who were attending the Medical Outpatient Department and who were in steady state were selected into the study at the University College Hospital, Ibadan, Nigeria between May 1996 and September 1998. The haematological profile and clinical evaluation of the patients were assessed. The steady state full blood count and reticulocyte counts were recorded. Packed cell volume was estimated as the spun haematocrit and the mean cell haemoglobin concentration was calculated from this value. Foetal haemoglobin was estimated by alkaline denaturation. Steady state was defined as the absence of acute illness or any chronic condition likely to influence the haematological profile. Systemic review was carried out on each patient. The spleen and liver sizes were measured.

Educational level and occupation of patients and their parents were noted. Age at menarche was noted for the females and age at noticeable secondary sexual characteristics was noted for the males. Psychological assessment of their concept of the disease was noted, notably depression, dependence on opioids and sexuality.

Previous transfusion requirements, number of hospital admissions per year and painful crisis per year were noted.

Liver function tests, electrolyte, urea and creatinine were assessed. Past history of cerebrovascular accidents, occurrence of priapism, chronic leg ulcers and acute chest syndromes were noted.

Results

Of the 98 patients in the study, there were 75 females and 23 males, giving a male to female ratio of 1:3. Age range was 30 to 52 years, the oldest being a female.

All the 98 patients had had some form of formal Western education. Twenty seven (25.5%) had a primary school leaving certificate, 23 (23.5%) had secondary school education and 48 (49%) were professionals. The professionals were comprised of accountants, bankers, engineers, teachers, nurses and hospital maids. All the patients were aware of the disease and had a good knowledge of the disease outcome.

Nine female (12% of female) patients started their periods between 13 to 15 years of age, 35 (46.6%) after 15 to 17 years, 27 (36%) after 17 but before 20 years and two (2.6%) started after 20 years. Seven (7.1%) of the patients were observed to have chronic leg ulcers. The leg ulcers spanned a period of five to 12 years before the study. Eighty six (87.7%) patients had never had a blood transfusion, three patients had been transfused three times, four had two units and five had one unit each (Figure I). Seventy six (77.5%) patients have never been admitted to hospital, while 22 have been admitted for mild to moderate infarctive bone pain crisis. Two patients had been admitted for skin graft of chronic leg ulcers during the course of the study. Haematological parameters of the patients showed packed cell volume (PCV) in the 21 to 28% range with a median of 24%. Second and third quartiles are 23 and 25% respectively. Reticulocyte counts were 1.2 to 7% with a median of 4.5%. Haemoglobin F in 33 patients was greater than three to less than 5%. Thirty six had HbF greater than 5 % but less than 10%. Twenty three patients had HbF greater than 10% but less than 15%. Five patients had HbF level greater than 15%. One patient had a haemoglobin F level of 22% (Figure II).

Figure I:Transfusion in SCA patients above 30.

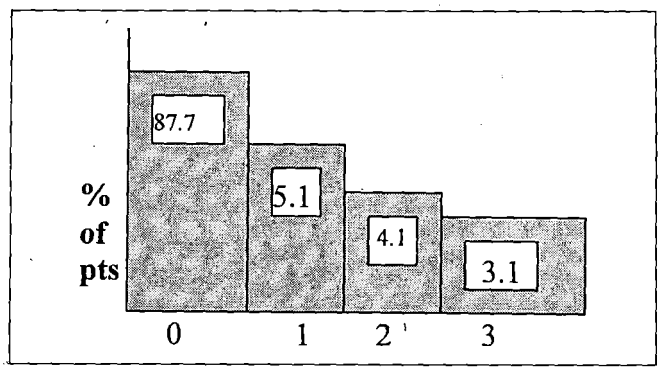
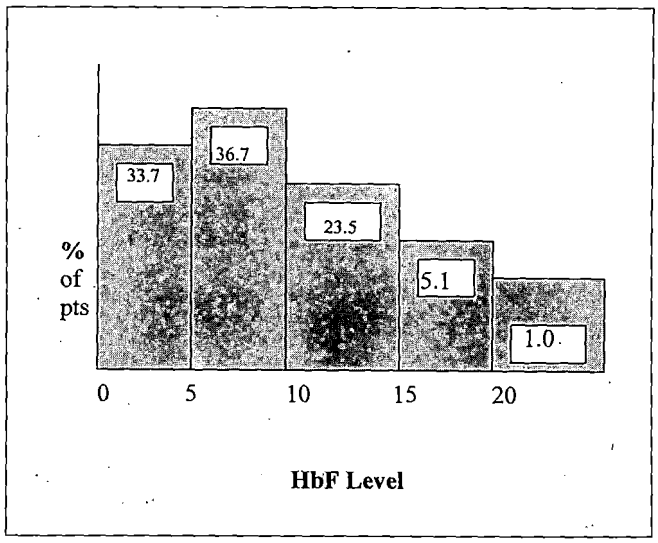


Figure II: Haemoglobin F HbF level in SC patients above 30 years.



Seventy patients (71.4%) had a normal range of urea and electrolytes. Three patients among the males have had priapism for which cavernospongiosal shunt procedure was carried out with consequent loss of penile erection. Five patients had a history of cerebro-vascular accidents which were managed conservatively and had shown no residual damage. Of the 75 females, 53 (70.6%) were married and 47 (62.6%) had been pregnant, and 23 had carried normal uncomplicated pregnancies.

It is worth noting that 34 patients (35%) had a mild to moderate degree of splenomegaly while 64 patients had no palpably enlarged spleen. All the 98 patients had mild to moderate hepatomegaly.

Discussion

Previous reports have observed a haemoglobin F level of 3 to 5% in this environment.[8] This trend was observed in only 33 (33.7%) patients in the present study. The 65 patients (66.3%) having HbF in the five to 22% range are unusual for this environment. These patients, however, have been alive for 30 or more years. The clinical complications, the degree of anaemia and the clinical severity have shown some degree of correspondence with the haemoglobin F level. For example the cohort of patients with haemoglobin F greater than 10% had no one with a history of cerebro-vascular accident, priapism or chronic leg ulcer. However, all have had a history of mild to moderate bone pain crisis and jaundice. Thus, the haemoglobin F level above 10% has not shown a protection against infarctive or haemolytic crises.

The 15 (15.3%) patients with features of gnathopathy, bossing of the forehead and some of the other features of the usual habitus of haemoglobin S were in the 3 to 5% HbF range. This observation is in agreement with Powars who concluded that a Haemoglobin F level above 10% reduces bone pain crisis and end organ damage. The Haemoglobin F level greater than 10% no doubt has some degree of protection against severe clinical complications in the sickle cell anaemia patient. All the patients had very few bone pain crises per year and had shown a good clinical severity score since diagnosis. In the study, a high percentage of patients had never been hospitalized nor had a blood transfusion for anaemia. The Haemoglobin F level however, did not show any significant difference between the females and males. The packed cell volume also did not exhibit any significant difference between the males and females. Though some age related complications are observed, the reticulocyte count, however showed no obvious relationship with level of haemoglobin F.

It is, however, noteworthy that all the patients had received some form of Western education. This may account for their heightened awareness of the nature of the disease and knowing the importance of hospital management and prompt medical consultation in time of any emergency. The only constraints might be finances or transport to the hospital. Eighty two per cent of the same cohort of patients admitted having parents especially mothers who are aware of the nature of their disease; mothers who are quite sympathetic and supportive. Only 14 (14%) reported that their mothers occasionally show frustration especially when they have to take care of more than one child in the family that has SCA. It is worth noting, however, that the location of the study could also have an influence on the relatively high number of (SCA) patients above 30 years. The easy accessibility to the teaching hospital, and ready availability of health care facilities would have facilitated the management of these patients and so avoided or prevented early death.

The results of the study strongly suggested that for the survival of SCA patients, educational background, awareness of the hospital treatment are major contributory factors. Where parents are aware of the complications of the disease, prompt medical care, compliance to drug therapy and regular follow up are the rule, thereby reducing the morbidity and mortality that might arise from the disorder. Also early diagnosis, improved medical care, better knowledge of preventable complications of SCA by the patients and their families have contributed to an increase in the life-span of these patients. It is also observed that selected health oriented and disciplined life styles,

stable or sedentary occupation, and good family support contribute to better health.

References

1. Fleming AF, Storey J, Molineauz L. Abnormal haemoglobin in the Sudan Savanna of Nigeria. Prevalence of haemoglobins and relationships between sickle cell trait, malaria and Surviv. *Ann Trop Med Parasitol* 1979;73:1161-2.
2. Muhammad S, Shurafa MS, Anaada S, Prasad AS, Donald L, Rucknagel DL, *et al.* Long survival in sickle cell naemia. *Am Journal of Haematol* 1982;12:357-65.
3. Felice AE, Mckie KM, Cleek MP. Effect of α thalassaemia 2 on the development changes of haematological values in children with sickle cell disease from Georgia. *Am J Haematol* 1987;25:389.
4. Fleming AF. Sickle cell disease. A handbook for the general clinician. Edinburgh: Churchill Livingstone. Chap 2. 22-32.
5. Sergeant GR (1982). Geography and the clinical picture of sickle cell disease. An overview. *Ann New York Academy Sciences* 1989;565:109-19.
6. Powars DR, Linda S, Chan-Walter A, Schroeder WA. The variable expression of sickle cell disease is genetically determined. *Semin Haematol* 1990;27(4):360-76.
7. Mears JG, Lachman HM, Labie D, Nagel RL. Alpha thalassaemia is related to prolonged survival in sickle cell anaemia. *Blood* 1983;62:286-90.
8. Charache S, Richardson SN. Prolonged survival of a patient with sickle cell anaemia. *Arch Intern Med* 1964;113:844-9.
9. Stricker VP, Kemp JA, Metts JC. Prolonged survival of a patient with sickle cell disease. *Am Pract* 1962;9:584-90.
10. Falusi AG, Esan GJF. Foetal haemoglobin in sickle cell anaemia in Nigerians. *Afr J Med Sci* 1989;18:145-9.